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Proximal muscle weakness and juvenile dermatomyositis

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Background: Juvenile Dermatomyositis (JDM) is a rare but serious systemic autoimmune condition of childhood primarily affecting proximal muscles and skin, characteristic findings include Gottron papules, a heliotrope rash, calcinosis cutis and symmetrical, proximal muscle weakness.

Case: A seven a year old presented with leg pains for 6 weeks with progressive proximal muscle weakness. She was unable to engage in her sporting activities and was noted to have increasing difficulty with climbing stair with strong family history of autoimmune condition. On examination she had pinkish discoloration cheeks and right metacarpal joints with proximal muscle weakness. Her routine blood tests, including connective tissue screen, rheumatoid factor, Vitamin D, myositis antibodies were all normal apart from her serum creatinine phosphokinase. She was started on 2 courses of methyl prednisolone followed by oral prednisolone and methotrexate subcutaneously. She gradually improved especially her skin disease with no new rash and her muscle strength improved with the help of physiotherapy. There is consensus about the diagnosis of JDM strongly supported by classic clinical and MRI findings. An initially aggressive approach with combination therapies as part of the early therapy of JDM may result in better long-term outcomes, including the possibility of less calcinosis, fewer corticosteroid side effects and a higher frequency of inactive disease, although these findings need to be confirmed in controlled studies and with long-term follow-up data.

Biography

Dipali Shah is an Acute and Ambulatory Pediatric Consultant at West Middlesex University Hospital, which is a part of Chelsea and Westminster NHS Foundation Trust. She is a Pediatric Assessment Unit Lead. She has participated in many national and international oral and poster presentations.

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